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Case report of partial empty sella syndrome: A challenging cocktail for the anesthesiologist

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Abstract
Background: Eventhough panhypopituitarism is a rare endocrine system disease, the clinical manifestation can vary from subclinical to life threatening myxoedema coma. It could be due to either primary pituitary dysfunction/stalk dysfunction/hypothalamic dysfunction.
A 32 year old young female who presented with bilateral hip joint pain was diagnosed to have bilateral avascular necrosis of femoral head. On ellicting a detailed clinical history, she underwent treatment for infertility for 7 long years which comprised of multiple ovulation induction agents and finally conceived following in utero insemination. Although the gestational and peripartum period were uneventful, she had agalactorrhea, a warning sign of impending hormonal dysfunction which didn't seek much attention. Five years down the lane, on evaluation for dyslipidemia, she was found to have hypothyroidism, hypocalcemia, low serum calcitriol levels. A detailed brain imaging studies showed partially emptysella with small remnant pituitary tissue and was started on treatment as a case of panhypopituitarism.

Keywords: Partial empty sella syndrome, preop optimization, combined spinal epidural anaesthesia, bilateral avascular necrosis of femur, Bone marrow aspiration and injection, Epidural analgesia

Introduction
Empty sella syndrome is a condition where pituitary gland shrinks making the sella turcica to look empty. Partial empty sella is suggestive that some of the pituitary gland will be visible as remnants on brain imaging study like MRI scan. Partial empty sella syndrome is a rare condition leading to central hypogonadism and female infertility. Incidence of empty sella is 0.5%and vast majority of 85% are women. The exact cause can be congenital or acquired like tumor, surgery, radiation leading to weakness of sellar diaghragm.
Here we are reporting a case of partial empty sella syndrome associated with Panhypopituitarism on multiple hormonal supplements for bilateral core decompression and bone marrow aspiration of avascular necrosis of femoral head under combined spinal epidural anaesthesia.

Case report
A 32 year old female presented to preanaesthesia clinic with known history of partial empty sella syndrome with ongoing hormonal supplementation for the past one year. Initial warning symptoms were cold intolerance, dyslipidemia, failure to conceive spontaneously which reslted in infertility treatment, Agalactorrhea following delivery. No family history of same disorder.
On examination – she weighed 73 kg, with a height of 172 cm/BMI 24.74,with coarse violaceous striaed skin on forearm.
Heart rate -86/min, normal sinus rhythm, normal volume and character.
Blood pressure -130/76mmhg on left arm in sitting position.
Systemic examination of cardiovascular system, central nervous system, respiratory system, gastrointestinal and genitourinary system were within normal limits.
Airway Assessment after COVID RTPCR - Revealed mallampatti class 2, Normal thyromental distance, adequate and painless neck movements.
Spine examination – Painless and nontender revealed a very narrow interspinous space without kyphoscoliosis, no neurological deficits.

Hence we anticipated difficult intravenous access and difficult regional anaesthesia.

Routine investigation

Complete blood counts, differential counts, Coagulation parameters, renal and liver function tests were within normal limits.Although there was fluctuating blood sugar levels which is not uncommon in panhypopituitarism on steroid replacement therapy.

Chest X-RAY (PA view) was within normal limits, E.C.G - normal sinus rhythm, no ST-T wave changes,

Metabolic equivalents - >5 METS

Hormonal assays preop after optimization with hormonal supplements

T3 - 85ng/dl(normal 80 -200ng/dl), T4 – 6 microgram/dl (normal 5-12mcgm/dl), TSH- 2.17 uIU/ml(normal 0.27 - 4.20Uiu/ML), Ft3 - 2 picogram/ml(normal2.8-7.1pgram/ml), Ft4 - 0.92ng/dl (normal 0.93-2 ng/dl), Prolactin -2.81 ng/ml(nonpregnant normal <25ng/ml)

LH - 1.3IU/L (normal2-9IU/L), FSH – 4.15 (normal 3- 10 IU/L), Cortisol - 5.07 microgram/dl Alkaline phosphatase - 88IU/L(normal 44-147 IU/L), Vitamin D3 -33(normal 30-80 ng/ml), Serum calcium (total) -9(normal 8.6-10.3 mg/dl), Parathyroid hormone – 26.8(normal11-51 pg/ml))

Lipid profile - total cholesterol -228 mg/dl (normal <180mg/dl), Triglycerides – 90mg/dl (normal <150 mg/dl)HDL – 77 mg/dl (normal 30 -95 mg/dl), LDL – 153 mg/dl(normal <100mg/dl), LDL/HDL – 1.99(normal <3.5)

Brain imaging study /MRI – Partial empty sella syndrome with remnants of pituitary tissues with normal brain parenchyma.

DEXA Scan –Revealed no features suggestive of osteoporosis.

Preop Optimization: T.Thyronorm 88microgram once daily, T.Hydrocortisone10mg TID, T.Calcium, T.Vitamin D3, T. Rosuvastatin 10mg, T.Ethinylestradiol and desogestrel (which was stopped five days prior to surgery day).

Anaesthetic Management

After adequate preop optimization and with adequate blood products arranged, a detailed written informed consent was obtained for the planned anaesthesia procedure of combined spinal epidural anaesthesia along with a backup plan for general anaesthesia in view of anticipated difficult regional anaesthesia access. On the preop day patient received her regular medications along with T.Pantoprazole 40mg and was allied of anxiety with T.Alprazolam 0.25 mg, and N.P.O was maintained from 10pm.

On the day of surgery after preparation of OT, checking anaesthesia machine with emergency resuscitation drugs, intravenous fluids, infusion pump, difficult regional anaesthesia equipments and equipment for general anaesthesia patient was shifted to OT after administering morning dose of T.Thyronorm 88 microgram, T.Alprazolam 0.25mg, T.pantoprazole 40mg, Injection Hydrocortisone 100mg (one hour before shifting to OT). Morning asymptomatic episode of low blood sugar of 76mg/dl was managed with intravenous DNS.

On receiving inside OT: Monitors for ECG, Noninvasive Blood pressure, pulse oximetry, temperature probe were connected. Two wide bore cannulas were established under strict asepsis under local anaesthesia and was preloaded with 10ml/kg of balanced salt solution. In view of anticipated difficult regional anaesthesia access sitting position was opted for combined spinal spidural anaesthesia procedure. Under strict asepsis under local anaesthesia with 2%plain lignocaine was infiltrated in L3-L4 Space for epidural needle placement. In L3-L4 space by a single attempt 18 G Tuohy needle was inserted, epidural space was identified by loss of resistance to air technique as well as by hanging drop technique and epidural catheter was threaded and fixed at 10 cm from skin following which a test dose of 2ml 2%lignocaine with 1:2lakh adrenaline was given which revealed negative for intravascular/intrathecal route.

Under strict asepsis, local anaesthesia with 2ml 2%plain lignocaine was instilled into L4L5 space for spinal anaesthesia. In single attempt 25 G Quincke‘s babcock spinal needle was inserted, following a clear CSF flow 2.2ml 0f 0.5 % heavy Bupivacaine was given. Sensory block till T10 level was confirmed in supine position with stable vitals. Intraoperatively epidural topup with 3ml 2% Lignocaine with adrenaline and 3ml of 0.5% bupivacaine given followed by epidural infusion with 0.5%bupivacaine at 4ml/hr. Intraoperative period Injection hydrocortisone 100mg was repeated along with two blood glucose monitoring. Judicious intravenous fluids were used with hourly monitoring of urineoutput. Surgery completed uneventful. Postoperatively strict blood glucose monitoring and judicious intravenous fluids advised as panhypopituitarism is prone for diabetes insipidus as well. Postoperative analgesia was with epidural anaesthesia with 0.2%ropivacaine at 3 to 4 ml /hour and supplemented with Epidural buprenorphine at 150 microgram at twelve hourly interval. Postoperative period was uneventful with a very good patient satisfaction in view of postoppain relief, injection hydrocortisone 100mg was continued 6th hourly along with routine hormonal supplements, epidural catheter was removed with tip intact on second postop day under strict asepsis with normal local area and was discharged on fifth postopday with no neurological deficits.

Discussion

Partial empty sella refers to the radiographic appearance of enlarged or deformed sella turcica that is partially filled with cerebrospinal fluid. When it’s due to a developmental defect and accompanied by symptoms of visual disturbance or endocrine abnormalities it’s termed as primary empty sella syndrome. Secondary empty sella syndrome occurs following radiation therapy, tumor, surgery, postpartum hemorrhage leading to sheehan‘s syndrome.

Our patient had hypothyroidism, hypertriglyceridemia, hypogonadism with one year period of amenorrhea, hypocortisolism, vitamin C and D deficiency optimized with hormonal supplements.

MRI is the gold standard modality to confirm the diagnosis of empty sella. There is no definite treatment policy in children but when accompanied with endocrine dysfunction hormone replacement therapy of target gland is done.

Conclusion

Patients with panhypopituitarism for anaesthesia is a challenge for the anaesthesiologist. Hence an evidence based and updated team of anaesthesiologists role becomes mandatory for managing such a case along with good

communication with operating team as well.Significant preoperative systemic manifestations and systemic diseases secondary to panhypopituitarism should be optimized prior to surgery to avoid intraoperative catastrophes that may even challenge patient ‘s life. Always it’s a team work that makes a dream work and follow safe anaesthesia.

Conflict of interest: There is no conflict of interest from any authors.

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